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The 22nd Annual Scientific Meeting of Indonesian Neurosurgical Society (INS)
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The 2nd International Fujita Bantane Interim Meeting of Neurosurgery



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XX ABSTRACT BOOK

SS 20 – PEDIATRIC

NEUROSURGICAL ASPECT IN SYNDROMIC CRANIOSYNOSTOSIS

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Craniosynostosis is a congenital abnormality in the skull, caused by premature fusion of one or more sutures. The prevalence of craniosynostosis ranges from 3.1 to 6.06 per 10,000 births, 9% of which are syndromic craniosynostosis. Skull growth only occurs in normal sutures, if cranial growth is very limited, an increase in ICP can occur. Direct monitoring of ICP for at least 24 hours can help diagnose and make decision processes.

Craniosynostosis is a complex disorder and management requires coordinated effort from a craniofacial (CF) multidisciplinary team. Initial management is the evaluation of CF team members to determine acute intervention, elective or just follow up. Acute neurosurgical intervention if an increase in ICP and visual impairment is found, if there is no acute action plan, an elective surgical plan is made, in the form of calvaria expansion to reduce ICP and FOA to enlarge the orbital cavity in a certain age period. If there is no elective intervention plan, the patient is monitored to anticipate changes that lead to reevaluation of the management plan.

Keyword: craniosynostosis, multidisciplinary CF team, acute intervention, elective intervention, follow-up.

168 ABSTRACT BOOK

WELCOME MESSAGE

Dear Friends,

It is our great pleasure to invite you to The 5th WFNS Spine Committee Biennial Conference of WFNS which will be held at Bali, Indonesia between October 25th - 27th, 2018.

WFNS scientific committees try to contribute to the education and progress of sub-disciplines of neurosurgery. Spine surgery is getting a high interest and Spine Committee Symposia every two years are the largest activity of the committee. I am happy to invite you to Bali, Indonesia to endorse activities in this part of the world. This meeting will be in conjunction with the Annual Meeting of Indonesian Neurological Society, Asian Epilepsy Surgery Congress. On October 25, a one-day cadaver dissection course will be held in Surabaya.

The meeting aims to reach a large number of audience, thus contribute to the spine education in this area more effectively. There will be "intense", and full of excellent lectures from prominent experts, results of implementation of new procedures, case discussions, debate sessions, video demonstrations, and workshops from industry.

The location of our congress is Bali island, one of the most beautiful and exotic place of the world. We really hope that it will endow us with many precious and long-lasting memories to cherish.

We look forward to seeing you in Bali in October 2018

Co-chairman of the WFNS Spine Committee.



Mehmet Zileli



Michael G. Fehlings



Daniel J. Hoh

ABSTRACT BOOK XXI

SS 20 – PEDIATRIC

CHANGES OF SUBVENTRICULAR ZONE NEURAL STEM CELLS IN HYDROCEPHALUS: AN EXPERIMENTAL ANIMAL MODEL

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ABSTRACT:
Background: Hydrocephalus, due to any cause, is still prevalence among pediatric group. It interferes with the developmental and growth of the child that harbor this condition. At present, the prevalent treatment is to perform a ventriculo-peritoneal shunt, which, for reasons that are not clear, is sometimes ineffective. We rely on the measurement of the cortical mantle as the primary predictors for the treatment result. The subventricular zone (SVZ) of the lateral ventricles has been known as the primary site of neurogenesis. Neural stem cells (NSCs) reside in the SVZ as a response of injury or stimuli, and can both differentiate into neurons and glial cells. However, only a few systematic studies of the role of NSCs in hydrocephalus have been reported. In a rat model of communicating hydrocephalus, we recently showed that hydrocephalus caused the ventricular system to expand over time and affect the SVZ.
Objective: To analyze the changes of SVZ in hydrocephalic animal model and its contributing factors.

Method: The experimental animal study design was employed using Sprague-Dawley rat of 10-12 weeks of age. Kainic-induced hydrocephalus protocol was applied to the rat model as described by Khan et al. The rat was divided into 4 groups: one normal group, 3 hydrocephalic groups (7 day, 14 day, 21 day). Hematoxylin-eosin and immunohistochemistry staining for beta-catenin, Ki-67, 4-HNE were studied.

Results: HE-staining shows changes in SVZ thickness among groups (1970.01 mm in normal group vs 1363.71 in 21-day-hydrocephalus group). Staining for Ki-67 shows that the proliferative activity in SVZ was depressed in 7-day-hydrocephalus (4.05) and started to incline on 21-day-hydrocephalus (7.87). The high 4-HNE score in hydrocephalic groups shows a high by-product of ischemic-reperfusion (8.95 in day 7 and 10.30 in day 21).

Conclusion: Traditionally cortical mantle thickness still can be used as reliable predictor. However, the changes in SVZ should be put into account as it also predicts the future of cellular recovery after shunting.

ABSTRACT BOOK 169

Aspek Bedah Saraf dari Kraniostenosis Sindromik

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Abstract

Craniostenosis is a congenital abnormality in the skull, caused by premature fusion of one or more sutures. The prevalence of craniostenosis ranges from 3.1 to 6.06 per 10,000 births, 9% of which are syndromic craniostenosis. Skull growth only occurs in normal sutures, if cranial growth is very limited, an increase in ICP can occur. Direct monitoring of ICP for at least 24 hours can help diagnose and make decision processes.

Craniostenosis is a complex disorder and management requires coordinated effort from a craniofacial (CF) multidisciplinary team. Initial management is the evaluation of CF team members to determine acute intervention, elective or just follow up. Acute neurosurgical intervention if an increase in ICP and visual impairment is found. If there is no acute action plan, an elective surgical plan is made, in the form of calvaria expansion to reduce ICP and frontoorbital advancement (FOA) to enlarge the orbital cavity in a certain age period. If there is no elective intervention plan, the patient is monitored to anticipate changes that lead to reevaluation of the management plan.

Keyword: craniostenosis, multidisciplinary CF team, acute intervention, elective intervention, follow-up.

Pengantar

Definisi

Kraniostenosis adalah gangguan pertumbuhan kalvaria akibat fusi prematur dari satu atau lebih sutura. Fusi prematur ini menyebabkan pertumbuhan abnormal kalvaria karena ketidakmampuan sutura untuk mengakomodasi pertumbuhan otak.

Epidemiologi dan genetika

Prevalensi berkisar dari 3,1-5,06 per 10.000 kelahiran (1–3). 84% pasien kraniostenosis datang dengan kraniostenosis *isolated*, 7% dengan gejala klinis tambahan dan 9% dengan kraniostenosis sindromik (3). Kraniostenosis sutura tunggal, lebih umum daripada multisutura. Tipe tersering adalah kraniostenosis sagital, yaitu 40-60% dari semua kasus (2–4). Untuk kraniostenosis multisutura, terbanyak adalah kraniostenosis bicoronal dan dapat muncul pada pasien sindromik atau non-sindromik (5). Kraniostenosis telah dijelaskan di lebih dari 150 sindrom yang berbeda (6,7).

Mutasi Fibroblast growth factor receptor (FGFR) paling sering dikaitkan dengan kraniostenosis, terutama tipe sindromik. Mutasi biasanya mengarah ke cacat genetik dominan autosom (8). Beberapa sindrom kraniofasial (CF) yang terkenal dengan mutasi FGFR termasuk Apert, Crouzon, Pfeiffer, dan Muenke. Sindrom lain, seperti Saethre-Chotzen dan Carpenter, terkait dengan mutasi non-FGFR.

Pertumbuhan tengkorak hanya terjadi pada sutura yang normal, sehingga menghasilkan bentuk kepala yang abnormal. Jika pertumbuhan kranial sangat terbatas, seperti terlihat pada kraniostenosis multisutura, dapat terjadi peningkatan TIK. Pasien dengan kraniostenosis tunggal cenderung mengalami peningkatan TIK daripada pasien dengan kraniostenosis multipel sutura dan / atau sindromik. Di antara pasien dengan multipel kraniostenosis, mekanisme multifaktorial yang mendasari peningkatan TIK termasuk disproporsi sefalokranial dan obstruksi aliran keluar vena. Pemantauan langsung TIK setidaknya selama 24 jam dapat membantu mendiagnosis dan proses pengambilan keputusan.

Kraniostenosis adalah gangguan yang kompleks dan manajemen memerlukan upaya terkoordinasi dari tim multidisiplin. Sindrom CF memberikan tantangan unik kepada tim CF, yang membutuhkan tingkat kewaspadaan yang konstan untuk memenuhi kebutuhan masalah yang kompleks ini. Dipandu oleh koordinator CF, spesialis bedah dan medis terlibat dalam perawatan pasien dengan pola pikir kualitas dan terus-menerus akan memperbaiki hasil. Tim CF multidisiplin yang terkoordinasi dengan baik adalah entitas yang menantang untuk diciptakan, membutuhkan banyak spesialis dengan tujuan bersama yang memiliki semangat dan ketekunan untuk terus berusaha memberikan perawatan yang lebih baik bagi pasien mereka.

Manajemen Awal

- Evaluasi untuk intervensi segera: setiap kasus dievaluasi secara individual, dengan tujuan mengantisipasi peningkatan TIK, gangguan visus dan saluran napas
- Jika tidak ada rencana tindakan akut, maka dibuat rencana bedah elektif, berupa ekspansi kalvaria untuk mengurangi TIK dan FOA untuk memperbesar rongga orbita.
- Menetapkan rencana pemantauan: jika tidak ada rencana intervensi elektif, maka penderita dipantau untukantisipasi perubahan yang mengarah pada reevaluasi rencana pengelolaan.
- Terapi adjuvan: pengobatan gangguan sindromik CF adalah bedah, tanpa terapi adjuvan.

Follow up

- Diikuti hingga dewasa: perlu follow up teratur dari aspek bedah plastik, bedah saraf dan oftalmologis, mulai saat terdiagnosis hingga usia awal dewasa. Follow up tahunan dilakukan pada pasien fase stabil, sedang pada fase awal pengobatan atau periode tidak stabil, lakukan lebih sering.

- Evaluasi radiologis dan rekaman TIK: jika curiga dengan naiknya TIK atau gangguan visus, lakukan ulangan CT scan dan / atau MRI dan jika perlu, lakukan pemantauan TIK.

Persiapan Operasi Kraniostenosis Sindromik

Indikasi Pembedahan

1. Peningkatan TIK: Kegagalan untuk segera mengenali TIK yang meningkat dan pertumbuhan otak yang terbatas karena fusi sutura akan menyebabkan hasil yang buruk. Penting untuk membuat keputusan awal tentang waktu dan tingkat intervensi bedah dini.
2. Pertumbuhan abnormal tengkorak yang progresif: operasi harus dilakukan sesegera mungkin bila ada hambatan pertumbuhan tengkorak, orbita terekspos atau obstruksi jalan napas berat.
3. Usia vs situasi klinis: Saat membuat keputusan operasi segera, pertimbangan klinis harus juga menakar risiko terkait operasi neonatal. Keputusan multidisiplin harus diambil pada aspek ini dan harus disesuaikan kasus demi kasus. Pada neonatus, selama bayi tidak menunjukkan tanda peningkatan TIK, obstruksi jalan napas, atau proptosis, tidak perlu terburu-buru untuk operasi segera.
4. Ketersediaan tim CF: Pasien kraniostenosis sindromik, harus dikelola oleh tim CF multidisiplin yang berpengalaman.

Persiapan Preoperatif

- Penilaian toleransi operasi: Setiap anak yang memerlukan pembedahan kranial untuk kraniostenosis sindromik akan menghadapi operasi besar setidaknya 4-6 jam dan kehilangan darah yang signifikan, oleh karena itu pra operasi harus dievaluasi kesehatan umum anak, status gizi, jantung dan pernapasan.
- Cross-match darah: Faktor darah dan pembekuan darah harus tersedia untuk memungkinkan penggantian volume darah jika diperlukan.
- Risiko infeksi saluran nafas: swab hidung dan tenggorokan pra operasi. Berikan antibiotik profilaksis secara rutin.

Pertimbangan Anestesi

- Anomali jalan napas: Pada anak-anak dengan saluran nafas atau *craniocervical junction* tidak normal, maka harus dipertimbangkan intubasi fiberoptik dan penundaan ekstubasi. Trakeostomi preoperatif mungkin diperlukan.
- Kehilangan darah: Tergantung pada preferensi tim, penggunaan asam traneksamat dan *cell saver* dapat dipertimbangkan untuk mengurangi kehilangan darah dan kebutuhan transfusi.
- Positioning: Pasien harus diposisikan secara hati-hati oleh ahli bedah dan ahli anestesi, dengan mempertimbangkan posisi kepala (prone atau supine), saluran napas, area tekanan kulit dan keratitis exposur. Tarsorafi mungkin diperlukan pada awal prosedur.

Implan

Sistem fiksasi tulang: untuk prosedur rekonstruksi tengkorak, diperlukan fiksasi tulang dan bervariasi sesuai dengan preferensi tim. Screw dan pelat yang dapat diserap dan tidak bisa diserap dapat digunakan. Tidak ada fiksasi khusus yang terbukti lebih unggul dan pengalaman individual ahli bedah, paling bernilai dalam menentukan pilihan fiksasi tengkorak.

Alat tambahan khusus

Craniotome dan drill: Peralatan kraniotomi standar diperlukan untuk rekonstruksi tengkorak.

Operasi untuk Kraniostenosis Sindromik

Tujuan pembedahan adalah untuk memperluas calvaria agar cukup ruang untuk pertumbuhan otak normal serta untuk menciptakan keselarasan simetris dan estetika. Tim CF berhadapan dengan sutura yang fusi, kepala yang tidak rata dan ruang orbital dangkal. Sebagian besar unit CF sekarang mengadopsi teknik bandeau fronto-orbital (teknik Marchac dan Renier) dengan dahi atau belakang kepala mengambang, penggunaan *barrel-stave* dan graft tulang *split-thickness*.

Sebagai aturan umum, prioritas diberikan pada level orbito-kranial, dan setelah *midface* atas dan tengkorak stabil, baru difikirkan rekonstruksi hidung, maksila dan mandibula.

Posisi Pasien

- Tarsorrhaphy atau pelindung mata lainnya: ini sangat penting, terutama pada anak-anak dengan bola yang menonjol.
- Anomali spinal: kelainan tulang servikal dan adanya malformasi Chiari memiliki dampak signifikan pada penentuan posisi pasien.
- Dalam kebanyakan kasus, anak diposisikan terlentang dengan kepala di head rest, yang memungkinkan reposisi selama operasi. Reposisi ulang secara berkala penting selama operasi panjang untuk mencegah luka tekan.
- Prone bila operasi sisi posterior mendominasi: Anak ditempatkan prone jika dilakukan ekspansi posterior atau remodeling calvaria lengkap. Pengangkatan kepala yang mengurangi tekanan kulit secara teratur selama beberapa detik diperlukan setiap 20 menit dalam posisi tengkurap, yang harus diatur waktunya oleh jam dan dicatat saat dilakukan.

Pendekatan Bedah

Insisi bikoronal: Insisi bikoronal zig-zag atau lurus dilakukan di belakang garis tengah dari telinga ke telinga dan jika insisi yang lebih rendah diperlukan, dibawa ke belakang telinga. Sayatan ini memungkinkan akses ke bagian depan dan belakang kalvaria dan dapat dilakukan reinsisi untuk prosedur berikutnya. Keuntungan dari sayatan zigzag adalah menyebabkan lebih sedikit scar dan rambut tidak akan menjadi bagian dari sayatan. Mencukur tidak diperlukan karena insisi dapat dilakukan di garis rambut yang terbuka.

Rekonstruksi anterior

Kulit diflap ke anterior untuk membuka orbital rim: Penutup kulit diangkat secara terpisah ke kedua orbital. Perikranium dibedah dalam lapisan terpisah sampai tulang orbital dan sutura zigomatik terpapar secara bilateral dan otot temporalis diangkat ke arah pangkalnya. Lapisan perikranial yang terpisah dapat dipanen jika

diperlukan untuk perbaikan dural dan dapat digunakan sebagai *overlay* vaskularisasi setelah panel kubah kalvaria diganti.

Nasion dan lateral orbital rim terbuka: margo superior orbital lebih lanjut dibedah sampai nasion dan ligamen kantal lateral terlihat.

Rekonstruksi posterior

Refleksi kulit kepala posterior: Insisi zigzag bicoronal yang sama dapat digunakan untuk rekonstruksi posterior. Tergantung pada prosedur yang direncanakan, jaringan kulit kepala dapat diflap kembali ke *craniocervical junction* jika diperlukan.

Intervensi

Sebelum kraniotomi dilakukan, tim membuat keputusan tentang penempatan flap tulang, yang kemudian ditandai dengan metilen biru atau pensil.

Rekonstruksi kalvaria anterior

- Kraniotomi bifrontal dengan pemanenan dahi baru: Pada rekonstruksi anterior, dahi baru, yaitu, template Marchac, ditinggikan lebih dulu. Kemudian sisa dahi yang asli dibuang.
- Osteotomi melintasi garis tengah dan atap orbita: Setelah retraksi lembut kedua lobus frontalis, osteotomi dilakukan di kedua atap orbital dan di depan cribriform plate.
- Potong arkus zygomatic pada sutura untuk mengambil bandeau orbita: arkus zygomatic dipotong pada sutura, tulang hidung pada sutura hidung, dan bandeau orbita kemudian dilepaskan.
- Remodeling bandeau orbital: bandeau orbital dibentuk secara individual.
- Ganti bandeau dan dahi: Bandeau dan dahi baru dimajukan simetris dan dilekatkan pada tulang skuamosal dan temporal dengan wire atau plate. Tindakan ini akan menciptakan ‘nasal step’ yang akan menghilang dengan cepat ketika anak tumbuh.

- Tutup defek tulang: defek tulang ditutup dengan sisa tulang yang dirajang (*barrel-stave*) dan dengan *split bone graft*. Fragmen tulang difiksasi dengan jahitan, dan hindari penggunaan wire berlebihan.
- Perbaiki robekan dural: setiap robekan dura harus benar-benar tertutup untuk mencegah terbentuknya akumulasi CSF di bawah tulang yang devascularisasi atau, untuk kebocoran dural besar, dapat terjadi growing fraktur di antara lempengan tulang yang digantikan. Penjahitan dural dilakukan dengan memperkuat jahitan dengan perikranium.

Rekonstruksi posterior

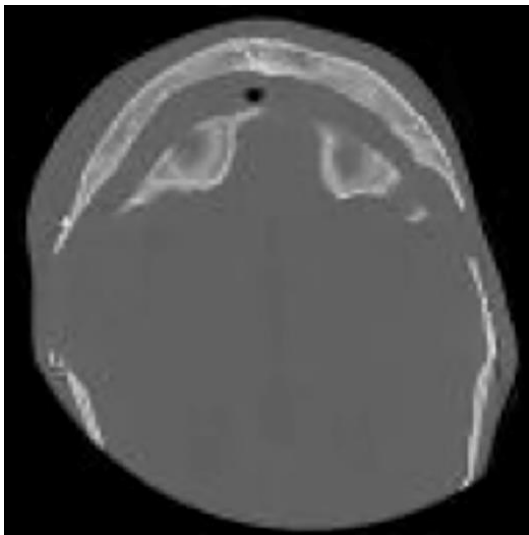
Rekonstruksi dan / atau ekspansi posterior diindikasikan jika area oksipital sangat terpengaruh, dengan penampilan yang rata dan pertumbuhan posterior yang sangat terbatas. Pada bayi yang tumbuh dengan TIK tinggi, ekspansi posterior memungkinkan pertumbuhan otak, sebelum waktu optimal untuk operasi frontoorbital definitif.

- Kraniotomi biparietal, termasuk garis tengah baru 'posterior': Pada rekonstruksi posterior, teknik bandeau digunakan untuk memperbesar dan membentuk kembali dua pertiga posterior dari kepala. Menggunakan teknik Marchac, fragmen "posterior" diambil dari bagian konveksitas yang lebih tinggi. Dari jendela craniotomy yang dihasilkan, kita dapat mendiseksi sutura sagital dan lambdoid. Osteotomi untuk mengangkat sisi posterior kepala tidak boleh lebih rendah dari asterion, untuk menghindari robeknya sinus.
- Reposisi bandeau dan garis tengah posterior baru: Bandeau baru difiksasi dengan pelat / sekrup atau kawat sesuai dengan preferensi ahli bedah.
- Mengisi sisa defek tulang: Setelah bagian belakang kepala yang baru telah dibentuk kembali, rekonstruksi selanjutnya dilakukan dengan cara yang mirip dengan rekonstruksi anterior.

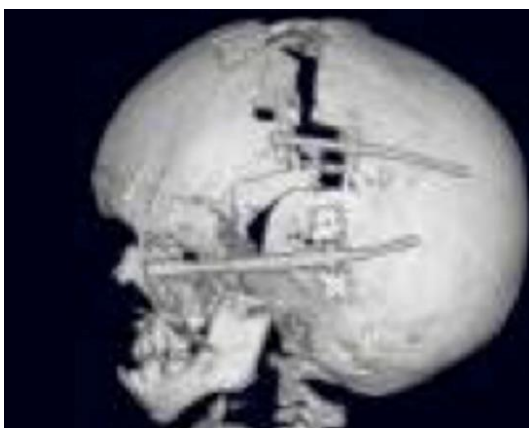
Teknik distraksi

Merupakan alternatif untuk teknik rekonstruksi kalvaria.

- Meletakkan distraktor setelah osteotomi: Osteotomi dibuat dan distraktor ditempatkan dengan poros mereka yang dibawa keluar melalui sayatan kulit yang terpisah.
- Distraksi bertahap selama berminggu-minggu: Kalvaria didistraksi dengan memutar poros distraktor untuk mencapai distraksi 1-mm setiap hari selama 3 minggu. Cara ini akan menghasilkan distraksi akhir 2 cm.
- Komplikasi potensial: Kelemahan teknik ini adalah tingginya insiden infeksi superfisial dan perlunya operasi kedua untuk mengangkat distraktor, dan potensi masalah penyembuhan luka. Akibatnya, teknik ini lebih banyak digunakan sebagai prosedur awal untuk perluasan kalvaria.



CT scan aksial dari distraksi anterior



3DCT: tampak distraktor terfiksir pada tengkorak posterior. Putaran $\frac{1}{4}$ dilakukan setiap hari selama 3 minggu untuk memajukan tengkorak dan wajah ke anterior.



Skull X-ray lateral dari distraksi posterior: Gambar menunjukkan distraktor dan pemisahan osteotomi.

Rekonstruksi Monobloc Anterior

Prosedur monobloc dilakukan setelah tumbuh gigi permanen di maksila, karena akan menghancurkan tunas gigi. Dalam prosedur monobloc, tulang frontal dan midface secara simultan dimajukan dalam dua segmen.

- Osteotomi dahi dan *orbital bandeau*
- Osteotomi Le Fort III: *Midface* dilepas dari basis kranii, enblok melalui Le Fort III osteotomy.
- Kombinasi ekspansi fronto-fasial: Kombinasi ekspansi fronto-fasial gabungan tidak hanya meningkatkan volume intrakranial tetapi juga mengoreksi eksoftalmos dengan memperdalam rongga orbita, memperbesar epifaring dan menormalkan oklusi gigi.
- RED Frame: ketika operasi monoblok dilakukan sebelum tengkorak pertumbuhan berakhir, sejumlah kelebihan tertentu diperlukan untuk memperhitungkan potensi pertumbuhan yang abnormal, tetapi kadang-kadang sulit untuk mencapai hasil yang diinginkan karena pembatasan jaringan lunak. Dalam kasus ini penggunaan perangkat gangguan internal atau eksternal telah menjadi opsi tambahan. Sedangkan gangguan internal terutama digunakan untuk kemajuan posterior, kerangka RED telah menjadi metode pilihan untuk fronto-facial monobloc kemajuan, karena mereka mampu mengontrol 3D yang lebih baik selama proses pengalihan.



3D rekonstruksi CT anak dengan REDframe



Rekonstruksi 3DCT kepala dengan RED frame

Spring

- Pada koreksi kraniostenosis sagital tunggal *spring* telah menjadi solusi lain untuk mempertahankan koreksi anatomi pada anak yang tumbuh dan berkembang.
- Ekspansi sutura lambdoid dengan *spring*, sebagai teknik alternatif untuk memperluas fossa kranial posterior pada pasien dengan kraniostenosis kompleks.

Timing Operasi untuk Kraniostenosis Sindromik

Apert Syndrome

- FOA pada umur 9–12 bulan: Saat sutura, fontanel, dan sinkondrosis menyatu, maka risiko hipertensi intrakranial meningkat. Pada saat itu, biasanya sekitar usia 9–12 bulan, merupakan indikasi FOA dini dengan remodeling kalvaria, karena ini akan menurunkan risiko peningkatan TIK. Perlu follow up teratur dan hati-hati, karena hipertensi intrakranial tetap dapat terjadi walaupun telah menjalani operasi. Jika ada tanda peningkatan TIK, maka perlu dekompresi dan remodeling lagi.
- *Midface advancement* pada masa kanak-kanak pertengahan: Usia untuk *midface advancement* bervariasi pada setiap pasien. Penatalaksanaan kelainan midface paling baik dilakukan pada pertengahan masa kanak-kanak, bersama dengan pembentukan kembali kalvaria anterior. Tindakan *midface advancement* terlalu awal beresiko membahayakan tunas gigi dan gigi permanen, seperti yang telah dibahas di atas. *Advancement* maksila terlalu dini, tidak mengarah pada normalisasi pertumbuhan sutura maksila.

Crouzon Syndrome

- FOA pada usia 9–12 bulan: Penatalaksanaan awal pada sindrom Crouzon biasanya membutuhkan FOA pada usia 9–12 bulan, kecuali anak mengalami eksoftalmos parah atau masalah pernapasan, maka koreksi segera.
- Ulangi FOA bila ada tanda peningkatan TIK: Anak diikuti pada interval teratur dan jika peningkatan TIK terjadi, maka ulangi dekompresi dan membentuk ulang kranial sesuai lokasi restriksi kalvaria. Selama prosedur ulang, risiko yang lebih tinggi dari robekan duramater dan kehilangan darah yang lebih besar harus diantisipasi.
- *Midface advancement* pada 5-7 tahun: Manajemen definitif dari deformitas *midface* melalui osteotomi monoblok, bipartisi wajah atau Le Fort III dilakukan antara usia 5-7 tahun. *Advancement* maksila lebih awal seperti yang dianjurkan oleh Tessier, tidak menormalisasi pertumbuhan maksila, sehingga dibutuhkan operasi elektif lebih lanjut dan perawatan ortodontik.

Pfeiffer Syndrome

Tahapan rekonstruksi sama seperti Crouzon, bergantung pada variasi individu.

Saethre-Chotzen Syndrome

- Koreksi kraniostenosis pada 8–12 bulan: Kraniostenosis pada anak dengan sindrom Saethre-Chotzen biasanya dikoreksi antara usia 8–12 bulan. Waktu serta sifat pembedahan disesuaikan dengan anak.
- Rencana pengobatan terkoordinasi untuk anomali terkait: Koreksi anomali terkait seperti langit-langit celah atau *webbed finger*, ditentukan dalam rencana perawatan terkoordinasi.

Follow up untuk Kraniostenosis Sindromik

Frekuensi Kontrol

Kunjungan rawat jalan pertama ke Klinik multidisiplin biasanya dilakukan 3 bulan, lalu 6-12 bulan. Klinik multidisiplin dengan layanan bedah plastik, bedah saraf, oftalmologis, psikologis dan genetik di tim inti. Saran ahli ENT mungkin diperlukan pada pasien dengan masalah saluran napas atau trakeostomi.

Frekuensi Foto

3DCT scan pascaoperasi dilakukan selama tahun pertama pasca operasi. Pemindaian lebih lanjut tergantung pada keadaan klinis, dengan timbulnya sakit kepala, gangguan visual, atau kekhawatiran perilaku menjadi indikasi untuk CT scan yang mendesak. MRI diindikasikan jika ada kekhawatiran klinis yang berkaitan dengan *craniocervical junction*. Selain itu juga diperlukan masukan dan penilaian tumbuh kembang dengan melibatkan dokter spesialis anak konsultan tumbuh kembang.

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